Pulmonary alveolar microlithiasis: review of Turkish reports

E S Ucan, A I Keyf, R Aydilek, Z Yalcin, S Sebit, M Kudu, U Ok

Gülhane Military Medical Faculty, Haydarpasa Training Hospital

Department of Chest Diseases, Medical School, Dokuz Eylül University, Izmir, Turkey

E S Ucan
A I Keyf
R Aydilek
Z Yalcin
S Sebit
M Kudu
U Ok

Reprint requests to:
Dr E S Ucan,
Türkay Cad.No 33,
Bornova, Izmir, Turkey

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Abstract

Pulmonary alveolar microlithiasis is a rare disorder, only 173 cases having been reported worldwide. Fifty two cases from Turkey are reported, 49 of which have previously been described only in Turkish publications. The mean age of the patients was 27 (SD 12) years, 34 were male, and 10 were symptomless. In 40 of the 52 cases diagnosis was confirmed histopathologically. Nineteen cases were diagnosed in siblings. This high rate suggests that pulmonary alveolar microlithiasis is a familial disease, which, though rare, is for unknown reasons most common in Turkey.

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Pulmonary alveolar microlithiasis is a rare idiopathic disease characterised by microliths in the alveoli, first described by Friedrich in 1856[1] and then by Harbitz in 1918.[2] O'Neill reported a family with pulmonary alveolar microlithiasis in 1967 and noted that the total number of recorded cases was 70.[3,4] Subsequently the number of published cases has risen to 173.[5-7] The first Turkish case was reported in 1962, and the total number of Turkish cases reached 52 this year.[8] As none of the Turkish cases, however, were included in any previous series we reviewed them and summarise their features here.[3,4,6,7]

Table 1 Characteristics of pulmonary alveolar microlithiasis cases in Turkey

Reference	*No of cases*	*Age (y) and sex*	*Family history*	*Diagnostic procedure*
Eber[6,24]	1	20M	−	OLB
Tezok[9]	1	28M	−	CNB
Alpaslan[10]	1	20M	−	CNB
Üner[11]	1	16M	−	CNB
Vidinel[12,13]	3	24F, 30F, 42M	+, +, −	XR, CNB, OLB
Gerelioglu[14]	1	21M	−	OLB
Özdogan[15]	1	22M	NA	OLB
Karasu[16]	1	18F	−	CNB
Özhan[17]	1	38F	−	CNB
Yücel[18,24]	1	21M	NA	OLB
Cakirca[19,20,24]	1	33M	−	CNB
Artvinli[21]	1	20M	−	OLB
Enacar[22,23]	6	10F, 12F, 15M	+, +, +	XR, XR, XR
		17M, 23F, 24M	−, +, +	XR, OLB, XR
Hacihanefioglu[24]	4	20M, 21M, 40M	NA, NA, NA	OLB, XR, OLB
		41M	NA	CNB
Akcan[25]	1	34F	NA	OLB
Kocyigit[26]	3	40F, 44F, 50M	−, +, +	OLB, OLB, OLB
Yenel[27]	2	28F, 38M	+, +	OLB, OLB
Kiliçaslan[28]	2	20M, 23M	NA, NA	CNB, BAL
Dogan[29]	2	21M, 26M	NA, NA	OLB, OLB
Balkanli[30]	1	68M	−	BAL
Türktas[31]	1	29M	NA	XR
Öztaskent[32]	1	35M	−	XR
Kanra[33]	3	4M, 34F, 20M	+, +, +	XR, OLB, XR
Celikel[34]	1	40F	−	BAL, TBB
Yüksel[35]	2	?M, 35F	+, +	OLB, XR
Camsari[36]	1	19M	−	BAL, TBB
Albayrak[37]	1	8M	−	OLB
Emri[38]	1	44F	−	OLB
Hazar[39]	4	30M, 46M, 17F,	+, −, +,	OLB, OLB, OLB
		8M	+	OLB
Uçan[40]	1	21M	−	OLB
Kamali[41]	1	35M	NA	OLB

OLB—open lung biopsy; CNB—cutting needle biopsy; XR—chest radiograph; TBB—transbronchial biopsy; BAL—bronchoalveolar lavage; NA—unreported.

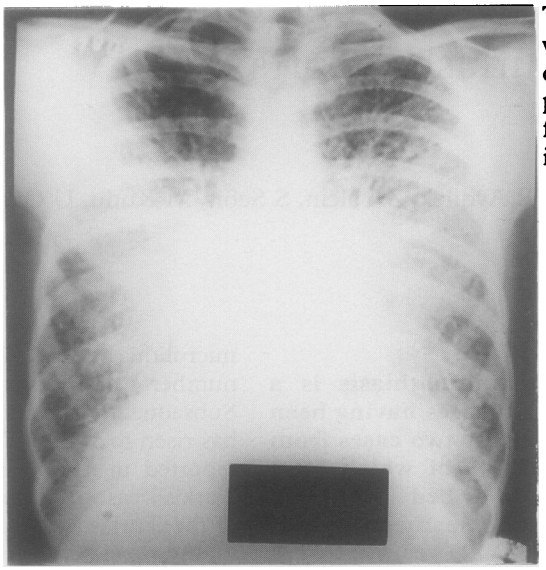
Cases

In all there were 34 male and 18 female patients with a mean age of 27·3 (SD 12) (range 4–68 years—see table 1). Ten of the 52 patients were symptomless but 35, 20, and five complained of dyspnoea, cough, and chest pain respectively. Physical examination revealed dullness and rales in 22, but no physical findings were noted in 22 of the patients. Cyanosis was noted in seven cases, clubbing in 12, and hepatomegaly in eight. Pectus excavatum and hypertrophic pulmonary osteoarthropathy were additional findings in two subjects.[30,38] The patients were from all the geographical regions of Turkey, though birthplaces were determined in only 23 cases.[9,22,23,26]

A family history was noted in 19 of the 39 patients. All the familial cases were siblings except for three who had a maternal aunt-nephew relationship. No family history was available in a quarter of the cases. Ten of the patients with a family history were female.

Nineteen of the patients had evidence of the disease on chest radiographs taken a mean of 3·9 years before the definitive diagnosis. All the radiographs showed the "sandstorm" appearances typical of pulmonary alveolar microlithiasis (figure). The initial diagnoses were sarcoidosis in one case, miliary tuberculosis in 13 cases and unknown in five. Only two with an initial diagnosis of miliary tuberculosis had acid fast bacilli in their sputum.

Although all of the 52 patients had a chest radiograph typical of pulmonary alveolar microlithiasis one patient also had bilateral apical pneumothoraces.[9] Bilateral diffuse calcification, pleural calcification, emphysematous bullae, and blebs were found in five other patients, who were examined by computed tomography.[34,36,38,40,41] Bone scintigraphy was also performed in seven patients and six had uptake of gallium in the lungs. Respiratory function tests were performed in 25 of the cases. The findings were normal in eight and the others all had a restrictive pattern. Arterial blood gas analysis in 15 patients



Chest radiograph showing typical sandstorm appearances of pulmonary alveolar microlithiasis

showed no hypoxaemia.

The diagnosis was made by open lung biopsy in 27 of the cases and by cutting needle biopsy, bronchoalveolar lavage, and transbronchial biopsy in nine, four, and four respectively. Radiological findings alone were considered to be sufficient in 12. Bronchoalveolar lavage fluid showed microliths in four of six patients, and transbronchial biopsy provided adequate material for histological examination in two of three patients.[28 30 34-36 40]

Discussion

The mean age of our group was similar to that in the cases overall. Nine of our 52 patients were below the age of 18 (table 2).[42 43] The geographical distribution of the patients in Turkey was heterogeneous. Although pulmonary alveolar microlithiasis has been reported from all over the world, 52 (23%) of the total of 225 cases now reported come from Turkey.[3 6 7 45] In addition, some of the patients reported from foreign countries were ethnic Turks. We therefore believe that pulmonary alveolar microlithiasis is more prevalent among Turkish people.[46 47] Some of the patients reported from other countries had been exposed to certain dusts. None of our patients had an obvious history of such exposure.[5-7 48 49]

This review reports 52 cases of pulmonary alveolar microlithiasis from Turkey, 49 of which have previously been described only in Turkish publications. Where family histories were available nearly half were positive for the disease, confirming previous reports that pulmonary alveolar microlithiasis may be a familial disease and suggesting that the gene is most common in Turkey.[3-5 37 43 44 47 50]

Table 2 Age and sex distribution, presence of family history, and symptoms in patients reported worldwide and in our series

Reference	No of patients	Mean age (y)	Male (%)	Family history (%)	Symptomless (%)
Sosman[3]	45	31–50	46	56	70
O'Neill[44]	27	30–50	50	38	—
Kino[43]	51	5–9	—	61	—
Prakash[6]	8	23	62	0	62
Mascie-Taylor[45]	71	32	49	—	87
Volle[46]	40	—	57	52	—
Our series	52	27	65	48	19

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